Sygnatura: © Pol J Radiol, 2007; 72(4): 86-90

Polish Journal of Radiology
www.PolRadiol.com
CASE REPORT

Otrzymano: 2007.04.25
Zaakceptowano: 2007.09.30

Inflammatory pseudotumor of the occipital condyle imitating a malignant neoplasm – a case report

Katarzyna Sznajder[1], Stanisław Skrzelewski[2]

[1] Department of Imaging Diagnostics, Regional Medical Center, Opole, Poland
[2] Chair of Public Health in Bytom, Silesian Medical University in Katowice, Poland

Author's address: Department of Imaging Diagnostics, Regional Medical Center, al. Witosa 26, 45-418 Opole, Poland, e-mail: k_sznajder@interia.pl

Summary

Background: Inflammatory pseudotumor is a non-neoplastic process of unknown etiology characterized by proliferation of connective tissue with an inflammatory infiltrate. IPT most frequently arises in the orbit, but can also be found in the larynx, the paranasal sinus and rarely in the skull base.

Case report: We present the case of a 20-year-old patient with a 4-month history of headache and insomnia. Neurological examination showed limited head mobility and hypoglossal nerve dysfunction. The patient was afebrile and no abnormalities in blood tests were found. CT revealed the presence of a tumor mass destructing the right occipital condyle. MRI was performed and the mass was surgically removed. The histological diagnosis was non-specific chronic inflammatory granulation tissue.

Conclusions: Inflammatory pseudotumors can often mimic malignant neoplasms, especially in cases where bone destruction is observed. IPT of the occipital condyle is a rare but aggressive lesion that should be treated by surgical excision.

Key words: inflamatory pseudotumor (IPT) • malignant neoplasm • occipital condyle

PDF file: http://www.polradiol.com/fulltxt.php?ICID=510551

Background

The axio-atlanto-occipital junction is an important region for many reasons. It is a structure allowing a great range of movement, and at the same time providing protection of important and sensitive CNS structures, such as the medulla oblongata and the spinal cold. It consists of bony, cartilaginous and ligamentous elements. The principal task of differential diagnostics in case of pathologic masses located in the central skull base region is to detect malignant tumors. Some lesions inflammatory or infectious in character may mimic a neoplastic process. Inflammatory pseudotumors (IPTs) account for a considerable proportion of such changes. They are nonspecific, polymorphic inflammatory cell infiltrates, accompanied by various grades of fibro-vascular component, clinically manifesting as a tumor [5]. The lesions of this type are most often observed in the orbit [9], less frequently in the larynx, paranasal sinuses, the infratemporal fossa, or the cervical spine [10]. Sporadic occurrence of IPTs in the bony structures of the skull base has been reported. They may be invasive, causing bone destruction and signs of impaired function of the cranial nerves. In view of nonspecific symptomatology, both clinical and radiological, as well as histopathological, IPT is diagnosed by exclusion of processes of other etiology. The treatment involves steroid- and radiotherapy, as well as partial or radical resection. However, both the response to treatment and the prognosis remain uncertain [5].

The paper presents a case of a 20-year-old patient with an IPT of the occipital condyle.

Case report

A 20-year-old patient was seen in the Admissions Department of the Neuropsychiatric Hospital with aggravating pains in the nape of the neck and occipital region of 4 months' duration. In the week preceding the admission

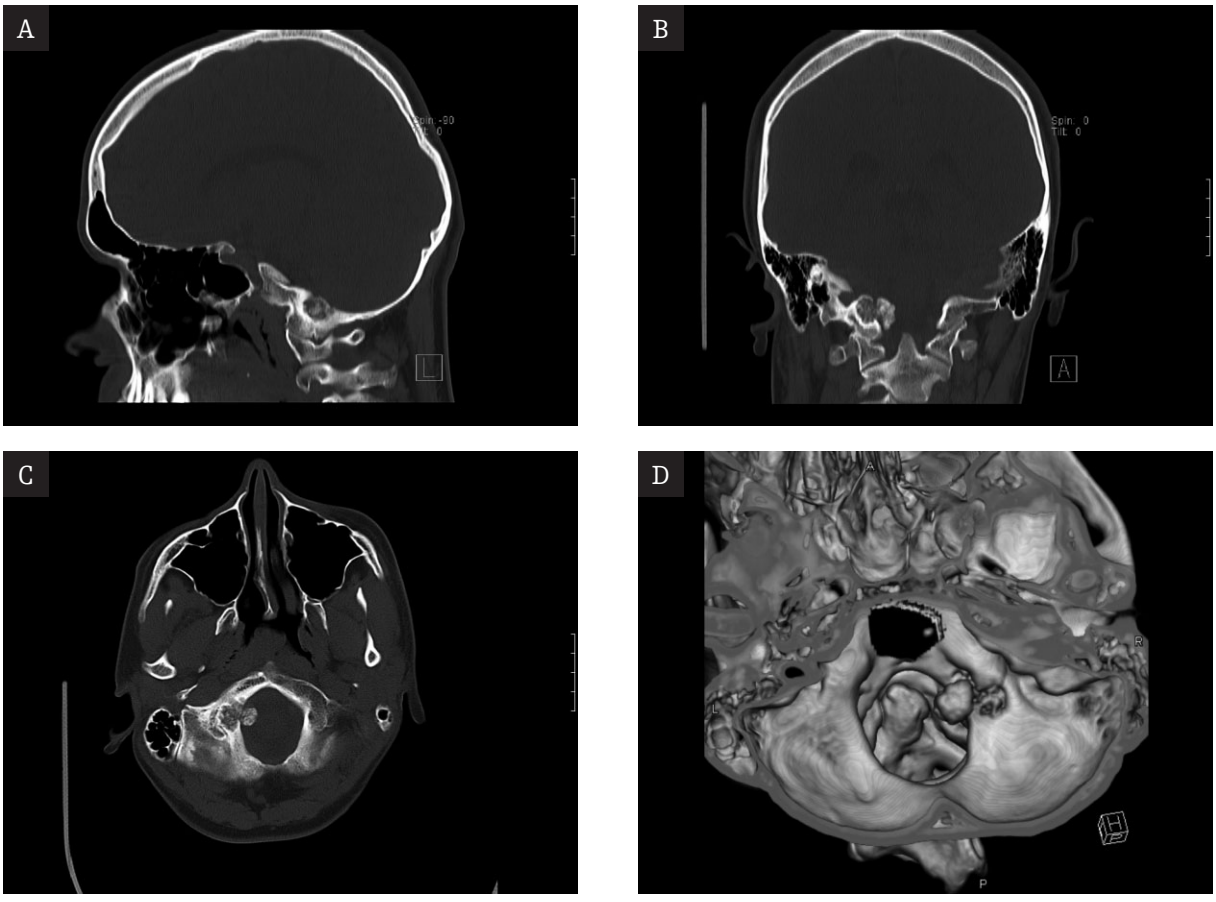


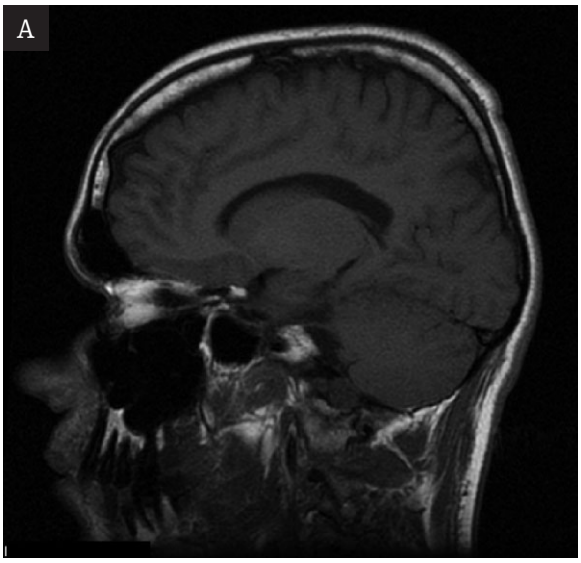
Figure 1. Multislice computer tomography of the head and cervical spine; bone window. A tumor mass causing bone destruction of the right occipital condyle. **A**) sagittal cross-section, **B**) coronal cross-section, **C**) axial cross-section, **D**) 3D reconstruction.

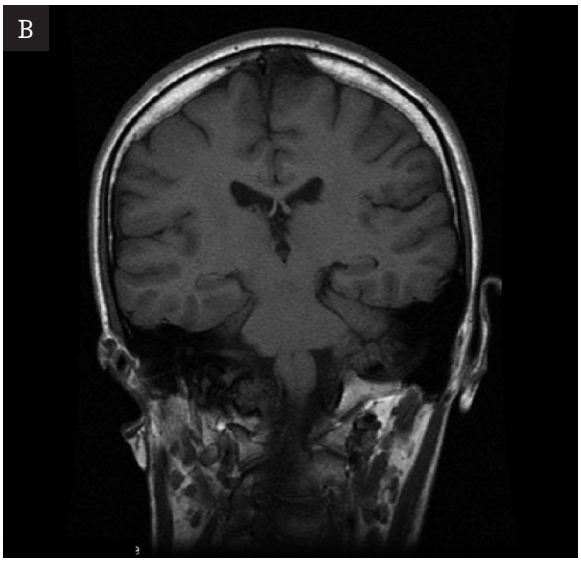
to hospital, the pain became unbearable, impairing normal functioning and sleep. The patient reported no history of any disorders and previous treatments. Physical examination revealed: the patient conscious, with logical contact, Kernig sign negative, nuchal rigidity difficult to assess because of pain of the cranio-cervical junction on bending the neck, limited possibility of turning the head to the left, increased paraspinal muscle tone in the cervical region. Additionally, slight left-sided tongue deviation was noted. Deep reflexes normal, without asymmetry. Muscular strength, superficial sensibility and Romberg test normal. Contrast-enhanced computed tomography of the head and non-contrast-enhanced CT of the cervical segment of the vertebral column was performed. A heterogeneous tumor-like structure of 20x15x17 mm dimensions, arising from the right occipital condyle, was visualized. The tumor extended into the foramen magnum, compressing the medulla oblongata and the right vertebral artery. The detected lesion was accompanied by destruction of the adjacent osseous elements (figure 1). The patient was referred to the Neurosurgery Department for surgical resection of the detected tumor.

On admission, left-sided head turn impairment was detected with no other physical and laboratory abnormalities. During the hospitalization period before the surgery, MRI of the head and CT of the cervical vertebral column for neuronavigation purposes. Suboccipital craniectomy and C1 laminectomy with redical resection of the lesion was performed. The postoperative course was uncomplicated. Control contrast-enhanced CT, which confirmed the radical character of the surgery and excluded the presence of early local complications, was performed. The patient was discharged home in good general condition with recommendations for follow-up in the hospital Outpatient Department of Neurosurgery and oncologist's consultation after obtaining the results of histopathology of the excised lesion.

Magnetic resonance tomography of the head was performed in SE/T1, T2 and FLAIR sequences in the transverse planes, SE/T1, T2 in the sagittal planes, and in SE/T1 sequence in the frontal planes. Sequential contrast-enhanced MRI was performed after intravenous administration of 10 ml of paramagnetic contrast medium, in SE/T1 sequence in the transverse, sagittal and frontal planes, with slice thickness of 5 and 3mm.

Within the foramen magnum, on the right side, a heterogeneous, well-delineated area was visualized. In SE/T1 sequence, it was characterized by intermediate signal intensity (figure 2). In SE/T2 sequence, the signal intensity was heterogeneous, with the predominance of hypointensive ones (figure 3) and was markedly, although heterogeneously, enhanced after contrast administration (figure 4). The visualized structure was merged with the occipital bone and infiltrated the spongy substance. It caused





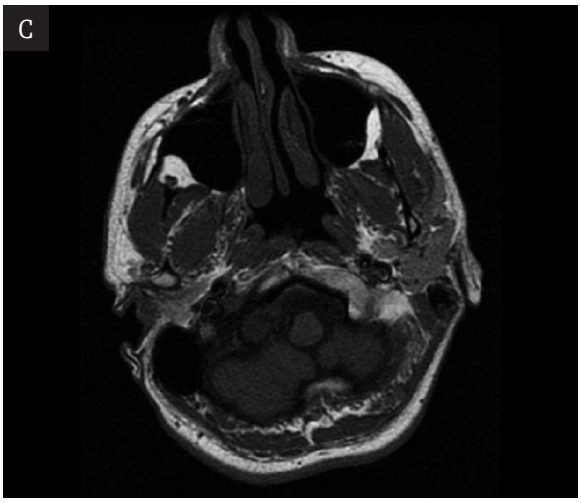


Figure 2. Intermediate intensity of the pathologic mass on T1-weighted images. It slightly shifts the medulla oblongata; **A**) sagittal cross-section, **B**) coronal cross-section, **C**) axial cross-section.

right-sided compression of the medulla oblongata, causing its deformation and slight displacement to the left. No signs of infiltration of the aforementioned anatomical structures of the nervous system were noted.

During the surgery, the presence of a solid, tumor-like lesion, not extending beyond the dura mater, was found. The lesion was excised together with a fragment of the occipital bone. The material submitted for histopathological investigations included 8 grayish-yellow, brittle specimens of the skull base tumor up to 0.6 cm in diameter. Microscopy revealed necrotizing-granulomatous inflammatory reaction. Elective staining did not reveal the presence of *Mycobacterium tuberculosis* or fungi. According to the histopatologist's suggestion, *Treponema pallidium* infection was excluded.

Discussion

Inflammatory pseudotumors of the head and neck form a heterogeneous group of idiopathic lesions, characterized by the presence of an inflammatory infiltrate with varied extent of fibrosis, localized most frequently within the orbit. The etiology of IPT remains unclear. The presence of chronic infections is considered, especially in the case of pulmonary IPTs, whose occurrence is associated with incomplete resolution of the inflammatory process. Typically, no destruction of osseous tissue is observed in the course of IPTs. However, it has been reported in the cases of tumors localized in the maxillary sinus and subtemporal fossa [7]. IPTs involving the skull base may give various symptoms, such as pain, compression of adjacent structures manifested by impairment of their function (cranial nerve paralysis, hypopituitarism). In case of tumors located within the foramen magnum, pains in the neck, paresthesias, contralateral hemiparesis can be observed. However, there are no specific clinical symptoms characteristic of the tumors located in that region. In the reported case, the symptoms included pains in the nuchal and occipital region, limited head mobility and slight abnormalities in the area innervated by the hypoglossal nerve. The diagnosis of an IPT can be established only on the basis of exclusion of other etiology of the process [1]. In the reported case, in view of many signs indicating the neoplastic character of the lesion and of vavorable anatomical location, it was decided to perform radical resection of the tumor. The histological classification of ITPs includes three subtypes, based on the observed ratio of cellular to fibrous elements. However, according to some authors, these proportions change with time in the course of the process, and the lesion itself, initially with predominantly cellular structure, at a later stage contains mainly fibrous tissue [4, 5]. The observed extent of fibrosclerotic reaction may also depend on the site of specimen collection [5]. In the presented case, this reaction was very limited. The most frequently observed tumors affecting the bony structures of the central skull base region include chordoma, chondrosarcoma, nasopharyngeal tumors infiltrating into that region, metastatic tumors and primary bone tumors. Such lesions are usually isointensive in T1-weighted images, and isolub hyperintensive in T2-weighted ones [6]. In the cases of skull base lesions of IPT type, their hypo- or isointensive

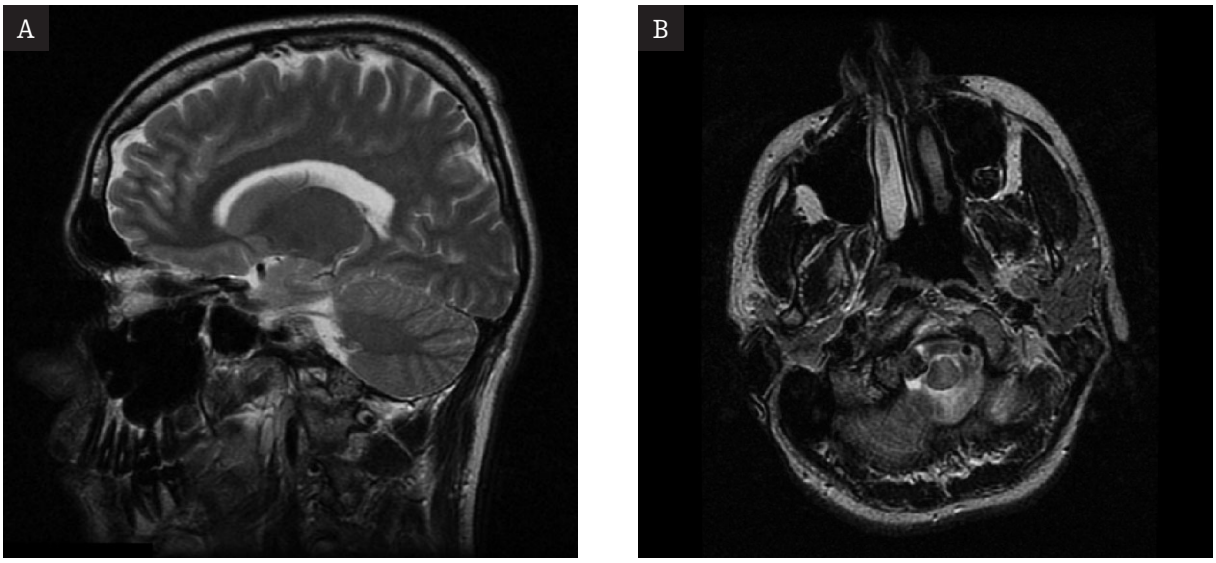


Figure 3. T2-weighted images show a low signal of the tumor; sagittal (**A**) and axial (**B**) cross-sections.

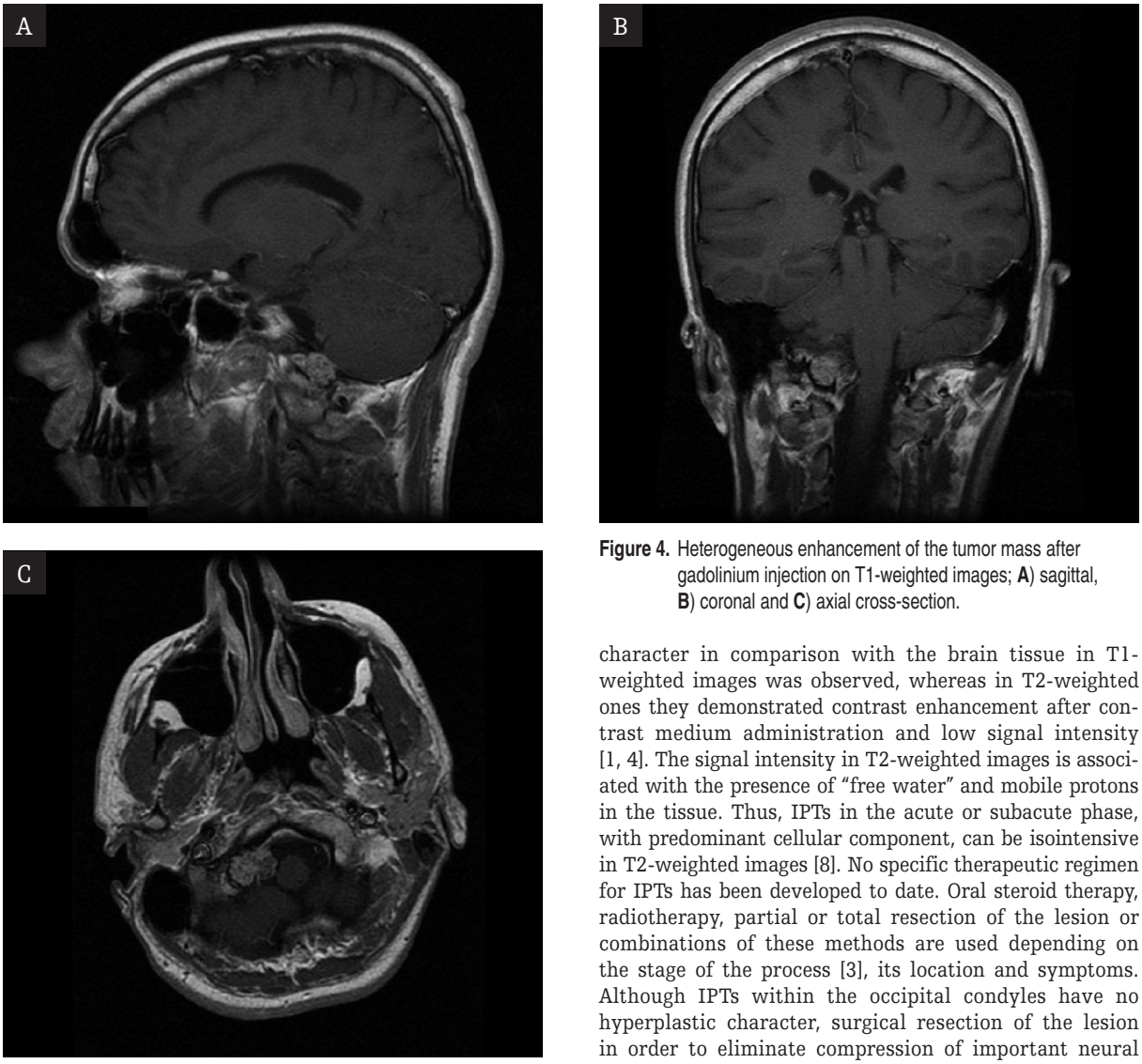


Figure 4. Heterogeneous enhancement of the tumor mass after gadolinium injection on T1-weighted images; **A**) sagittal, **B**) coronal and **C**) axial cross-section.

character in comparison with the brain tissue in T1-weighted images was observed, whereas in T2-weighted ones they demonstrated contrast enhancement after contrast medium administration and low signal intensity [1, 4]. The signal intensity in T2-weighted images is associated with the presence of "free water" and mobile protons in the tissue. Thus, IPTs in the acute or subacute phase, with predominant cellular component, can be isointensive in T2-weighted images [8]. No specific therapeutic regimen for IPTs has been developed to date. Oral steroid therapy, radiotherapy, partial or total resection of the lesion or combinations of these methods are used depending on the stage of the process [3], its location and symptoms. Although IPTs within the occipital condyles have no hyperplastic character, surgical resection of the lesion in order to eliminate compression of important neural

structures is the treatment of choice [2]. The aim of the therapy should be the inhibition of inflammatory activity and elimination of pain. Because of the presence of fibrosis in the lesion, as well as frequent cicatrization of the surgical wound, complete normalization of MRI results and reversal of neurological deficits may be unattainable [4].

Conclusions

Inflammatory pseudotumors can often mimic malignant neoplasms, especially in cases where bone destruction is observed. IPT of the occipital condyle is a rare but aggressive lesion that should be treated by surgical resection.

References:

1. Cho YS, Kim SM, Chung WH, Hong SH: Inflammatory pseudotumour involving the skull base and cervical spine. J Laryngol Otol, 2001; 115(7): 580–4.
2. Pallini R, Liverana L, Maria LL, et al.: Isolated inflammatory pseudotumor of the occipital condyle. Report of two cases. J Neurosurg, 2002; 97(2 Suppl): 248–51.
3. Mombaerts I, Schlingermann RO, Goldschmeding R, et al.: Are systemic corticosteroids useful in the management of orbital pseudotumors? Ophthalmology, 1996; 103: 521–7.
4. Sung MW, Kim KH, Lee CH, et al.: Fibrosing inflammatory pseudotumors of the central skull base. Laryngoscope, 1997; 107(12 Pt 1): 1651–5.
5. Batsakis JG, El-Naggar AK, Luna MA et al.: Inflammatory pseudotumors: What is it? How does it behave? Ann Otol Rhinol Laryngol, 1995; 104: 329–31.
6. Han MH, Chi JG, Kim MS et al.: Fibrosing inflammatory pseudotumors involving the skull base: MR and CT manifestations with histopathologic comparison. Am J Neuroradiol, 1996; 17(3): 515–21.
7. Sorn PM, Brandwein MS, Maldjian C et al.: Inflamatory pseudotumor of the maxillary sinus: CT and MR findings in six cases. Am J Roentgenol, 1994; 163:689–692.
8. Watanabe A, Ishii R, Okamura H, et al.: Magnetic resonance imaging of non-specific inflammatory granulation involving the skull base – two case reports. Neurol Med Chir (Tokyo), 1998; 38(2): 104–6.
9. Heersik B, Ridregues MR, Flanagan JC et Al.: Inflammatory pseudotumor of the orbit. Ann Ophthalmol, 1977; 9: 17–29.
10. Weisman RA, Osguthorpe JD: Pseudotumors of the head and neck masquerading as neoplasia. Laryngoscope 1988; 98: 610–614.